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Congenital duodenal obstruction due to duodenal atresia with preduodenal portal vein, annular pancreas, and intestinal malrotation associated with situs inversus abdominis: A case report



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ABSTRACT

The operations for congenital duodenal obstruction due to duodenal atresia with, annular pancreas still present a unique challenge. Of the operative techniques mentioned in the literature duodenoduodenostomy was the most frequently performed, however peculiar situations may arise that may warrant gastrojejunostomy. We report a case of duodenal obstruction due to duodenal atresia with preduodenal portal vein, and annular pancreas, co-existing with intestinal malrotation, a right sided stomach and spleen in which, the patient had gastrojejunostomy.

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Duodenal atresia is often associated with anomalies that include annular pancreas, cardiac anomalies, intestinal malrotation, situs inversus or splenic anomalies [1]. The associated duodenal atresia, annular pancreas, intestinal malrotation, is responsible for duodenal obstruction in a significant percent of patients with preduodenal portal vein (PDPV) [2]. However, an isolated cause of congenital duodenal obstruction from PDPV has been reported in only 4% of cases [3]. Here we report the clinical and anatomic findings of a 10-day-old boy with duodenal obstruction secondary to duodenal atresia, associated with an annular pancreas, intestinal malrotation, and PDPV.

1. Case report

Baby M.M was a 10-day-old boy who was delivered, per vaginam, to a family of non-consanguineous parents by a 20-year-old P₃⁰A₃ house wife after 39 weeks of uneventful gestation. The other siblings were doing fine. The mother had no history of use of recreational drugs, maternal diabetes during pregnancy and no history of congenital anomalies has been recorded in the family. The baby started vomiting on the 4th day of life. Vomiting was non-projectile,

bilious and post-prandial. The vomiting gradually became persistent with a frequency of 3–5 times/day. He also developed epigastric fullness 2 days into the onset of the vomiting. There was fever, but there was no jaundice and there was no problem when he passes urine. The baby's weight was 2.55 kg, axillary temperature –37.6 °C; he was not pale but was dehydrated. The apex beat was located in the fifth left intercostal space mid axillary line anteriorly. The heart sounds were clearly audible on the left side and no murmur was heard.

A plain abdominal X-ray showed the presence of a right-sided double bubble sign (Fig. 1). The remaining bowel loops were devoid of gas, which we assume it was a case of duodenal atresia with a probably wrongly labeled radiograph. The electrolytes were deranged.

The patient was rehydrated, electrolyte imbalance was corrected and he had a laparotomy through a supra umbilical transverse incision. Exploration of the abdomen revealed a right-sided stomach with a right-sided spleen (Fig. 2). The first part of the duodenum was edematous and dilated, and the portal vein was found anterior to it. The pancreas completely encircled the second part of the duodenum just distal to the preduodenal portal vein (Fig. 3). A portion of the body of pancreas extends to the third part of the duodenum. The intestines distal to the annular pancreas were collapsed, and the duodenal C-loop was incompletely formed which was also tethered by congenital adhesive bands (Ladd's

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bands); the cecum was on the left side of the abdominal cavity. The liver was in its normal position but the left lobe was bigger than the right lobe. A size 8F feeding tube passed intranasally could not go beyond the second part of the duodenum, suggestive of duodenal atresia. The congenital bands were divided and an appendicectomy was done. A loose over bridging gastrojejunostomy was done to bypass the obstructing annular pancreas and the pre-duodenal portal vein (Fig. 4). The wound was closed in a standard fashion.

The patient developed watery diarrhea on the third post operative day, which was aggressively managed with fluid and electrolyte replacement. Breast milk was gradually introduced via an NGT on the 8th day, which he tolerated well enough and was placed on direct breast feeding under supervision by the second post operative week. The patient was discharged on the 3rd post operative week. The patient had 8 weeks of uneventful post operative follow up before defaulting.

2. Discussion

Although duodenal atresia is the commonest cause of congenital duodenal obstruction, duodenal obstruction could still arise from other causes other than duodenal atresia [4]. In a literature review by Yi et al., [5] there were intestinal malrotation in 64%, pancreatic anomalies in 22% and situs inversus in 26% of cases with duodenal obstruction accompanied by PDPV. In our patient the obstruction was primarily due to the duodenal atresia with the co-existing annular pancreas. Persistence of vomiting after attempted feeding in our patient, and the fact that a size 8F feeding tube could not be passed beyond the second part of the duodenum intra-operatively suggests complete duodenal obstruction due to a duodenal atresia. Though duodenal obstruction due to PDPV as a single entity is uncommon, but it has been reported in the literature [6,7]. PDPV has also been found to cause duodenal obstruction that can manifest late in childhood or be accidentally diagnosed in adulthood [8,9]. Despite the low incidence of an isolated PDPV causing duodenal obstruction, it is still a potential hazard for hepatobiliary surgeons and intervention radiologist.

Embryologically, duodenal atresia is due to an error in recanalization of the duodenum; failure of the ventral pancreatic bud to rotate dorsally, makes it remain anterior to the duodenum thus



Fig. 2. Situs inversus of the stomach (a) right lobe of the liver, (b) the spleen.

fusing with the dorsal primordium to form a ring of pancreatic tissue (annular pancreas) around the duodenum; and finally, the persistence of the caudal anastomosis between the vitelline veins and the disappearance of the dorsal anastomosis with the caudal limb of the left vitelline vein predisposes to the formation of PDPV [10].

Persistence of the caudal anastomosis could explain the association of PDPV with malrotation, annular pancreas, and situs inversus but the reason as to why duodenal atresia and sometimes biliary atresia occur in association with PDPV still remains an assumption. Some studies have considered maternal diabetes, prematurity, exposure to retinoic acid and familial inheritance as possible risk factors for the development of duodenal atresia [3,11].

The presentation of bilious vomiting a few days after birth and the recurrence of the vomiting after an attempted feeding in this patient suggested a complete obstruction. Intraoperatively, the first part of the duodenum was edematous and dilated, and there was a ring of pancreas distal to the PDPV, and the body of the pancreas completely overlying the second part of the duodenum and extends to the third part of the duodenum. These finds precludes a conventional duodenoduodenostomy or duodenojejunostomy anterior to the portal vein [4]. Rather a gastrojejunostomy was performed after a modified Ladd's procedure (release of Ladd's bands and appendicectomy). The patient tolerated the procedure very well. We considered gastrojejunostomy a pertinent option in this patient for

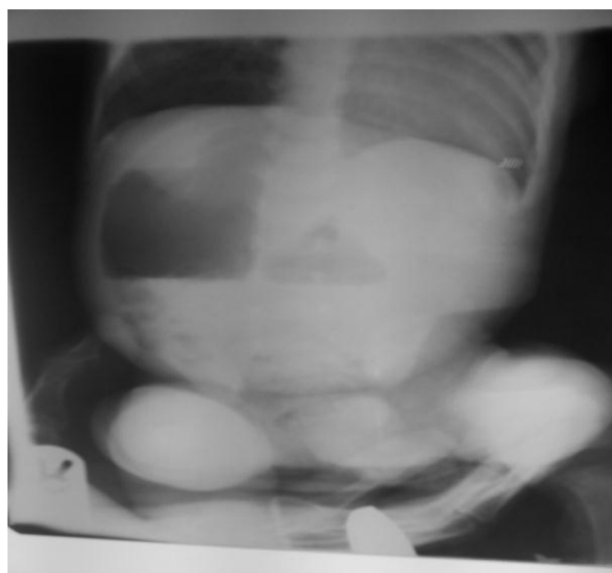


Fig. 1. Plain abdominal X-ray showing situs inversus with dilated stomach and first part of the duodenum (double bubble sign).

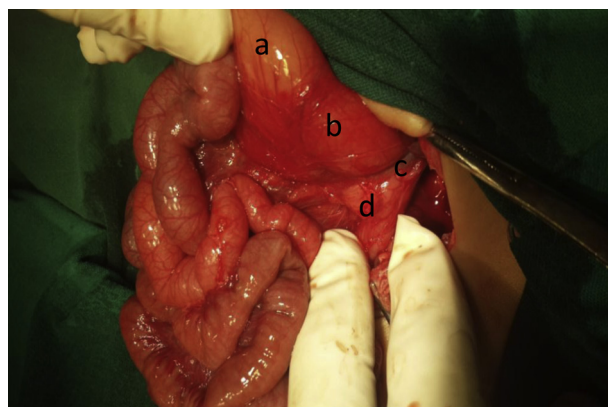


Fig. 3. Intra-operative findings: (a) right-sided dilated stomach, (b) first part of the duodenum, (c) Preduodenal portal vein (PDPV) crossing over the first part of the duodenum, and (d) ring of pancreatic tissue distal to the PDPV.

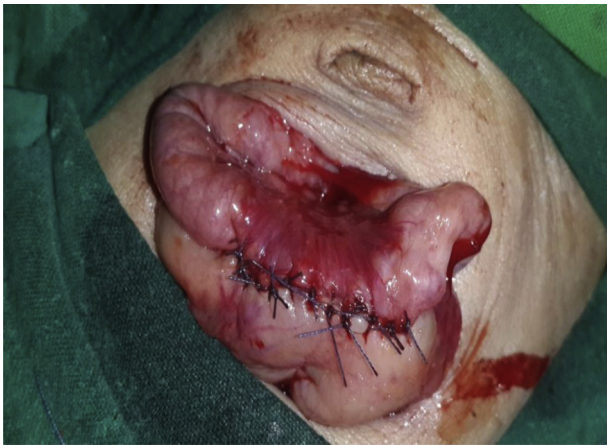


Fig. 4. Gastrojejunostomy distal to the PDPV and the annular pancreas.

the fact that, it allowed a loose over bridging in order not to obstruct the portal venous flow, secondly an undue tension may be exerted on the anastomosis and on the PDPV because of the extended overlying ring of the pancreas if we were to do duodenoduodenostomy or a duodenojejunostomy on the first part of the duodenum and thirdly, we avoided the dilated and edematous duodenum which can predispose the patient to prolonged ileus. Duodenoduodenostomy and duodenojejunostomy which were more physiological options have been proposed [12], but suffice it to mention that the anatomical defects determine the choice of procedure.

In conclusion, gastrojejunostomy is not a routine procedure for this condition, considering its non-physiological and also the risk of late complications such as a blind loop syndrome and marginal ulcer. But the procedure can still be considered in occasional cases of obstruction of the first and second parts of the duodenum.

Conflict of interest

Authors share no financial conflict of interest.

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